A unique approach in the management of vena caval thrombosis in a patient with Klippel-Trénaunay syndrome

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Vena caval thrombosis has posed a surgical therapeutic challenge for many years. Historically, spiral vein grafts and synthetic materials used as prostheses have had variable results. The use of the stent may serve as a more promising alternative when used in the capacity to relieve caval obstruction. A case is reported in which a young woman with Klippel-Trénaunay syndrome has exercise intolerance and associated hypotensive cardiovascular collapse caused by inferior vena caval thrombosis. Recanalization of her inferior vena cava was successfully achieved and subsequently maintained through the placement of two Wallstents across the lesion. Although most venous stenting procedures have thus far been used in the treatment of venous obstruction caused by malignancy, inferior vena cava stenting in this patient with inferior vena caval thrombosis and Klippel-Trénaunay syndrome suggests that venous stenting might offer an alternative therapeutic modality in treating a broader spectrum of occlusive venous disease. (J Vasc Surg 1997;26:155-9.)

Historically, thrombosis of the vena cava has posed a surgical therapeutic challenge. Early comparative studies that assessed patency rates for prosthetic and autologous grafts were disappointing.[1] Replacement of the vena cava with grafts is particularly hindered by its large diameter, low pressure, and low flow.[2,3] More recently, synthetic materials and prosthetic stents have provided the surgeon with alternative interventional management options. The stent, in particular, is a potentially promising device, in view of the poor results of previous materials.

In 1900, Klippel and Trénaunay[4] first described the pathologic condition that currently bears their names in the "Du Naevus Variqueux Osteohypertrophique." They described a syndrome characterized by a nevus on the affected extremity, varicose veins of the same limb, and hypertrophy of both soft and bony tissues of the affected extremity. Servelle,[5] in a review of 768 operations in patients with Klippel-Trénaunay syndrome, documented that the overwhelming majority of venous abnormalities occur in the lower extremity. Furthermore, limb elongation is invariably present, whereas the frequency of other associated symptoms and structural abnormalities can vary.[5] Anomalies of the deep veins of the involved extremity, including hypoplasia, atresia, aplasia, and valvular agenesis, can be dreaded components of this rare syndrome.[6] Often, large superficial varicosities develop as collateral conduits for the obstructed deep venous system.[5] Nevertheless, venous insufficiency may still ensue, resulting in stasis, severe edema, and other associated complications. The therapeutic dilemma of managing vena caval thrombosis combined with the rare and complicating condition of Klippel-Trénaunay syndrome posed a difficult and interesting clinical problem in a patient recently treated at our medical center.

CASE REPORT

The patient was a 27-year-old woman with a medical history significant for Klippel-Trénaunay syndrome, notable for a long history of venous abnormalities of the left lower extremity. She had a deep venous thrombosis of the left lower extremity, with a subsequent pulmonary embolus prompting the placement of a caval filter in October 1988.

She recently came to our medical center with symptoms of increased leg swelling with exercise and associated

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Fig. 1. Iliac venogram with thrombosis of the inferior vena cava. Note the anomalous left iliac venous system *(arrow)*.

Fig. 2. Reconstituted inferior vana cava. Note the Greenfield filter at the inferior portion *(arrow)*.

hypotensive cardiovascular collapse, relieved by leg elevation. The patient had many of the associated stigmata of Klippel-Trénaunay syndrome, including unilateral lower extremity hypertrophy, edema, and port-wine stains. A duplex ultrasound examination was performed and was difficult to interpret because of the patient's underlying venous anomalies. A physiologic workup involving upright cycle ergometry was performed that revealed that the patient had poor exercise tolerance, stopping after 4 minutes, with diminished cardiac preload caused by decreased venous return. A venogram revealed large anomalous venous drainage channels of the left lower extremity, consistent with Klippel-Trénaunay syndrome (Fig. 1). It further revealed thrombosis of the inferior vena cava at the level of the Greenfield filter as a cause of her diminished cardiac preload (Fig. 2; note the low placement of the Greenfield filter). An aortogram was also performed that effectively ruled out any arterial component to her vascular anomalies.

She then underwent a revascularization procedure of the vena cava to improve her venous return. To facilitate recanalization of the vena cava, the filter was removed by direct open surgical approach. A cavotomy was performed and the filter was excised. The organized thrombus was resected and the vena cava was repaired primarily. Under direct vision, the remainder of the vena cava was recanalized by balloon angioplasty. A Wallstent was subsequently placed in the area of the caval angioplasty under direct vision.

Before the operation, the patient had a lower extremity venous pressure of 27 cm of water, whereas the suprahepatic caval pressure measured 10 cm of water, thereby yielding a 17 cm of water pressure gradient. After deployment of the stent, there was equalization of venous pressures in the operating room. The patient was fully anticoagulated with heparin after surgery. The patient tolerated the procedure well, but a retroperitoneal hematoma and recurrent caval thrombosis subsequently developed, presumably because of the increased retroperitoneal pressure from the hematoma, the transient hypovolemic hypotension, or the technical misadventure with the stent (Fig. 3). The heparin was discontinued and the patient was returned to the operating room for evacuation of the hematoma. She tolerated this procedure and began treatment with heparin and warfarin on postoperative day 2. She was then discharged home from the hospital and continued treatment with warfarin, with plans for recanalization of the inferior vena cava in 3 weeks. After readmission, a percutaneous venogram was performed through a right femoral approach, demonstrating thrombosis of the stent. Balloon angioplasty was performed to recanalize the stent and the inferior vena cava. There was residual stenosis cephalad to the primary stent, and a second Wallstent was placed, overlapping the first. A final venogram was obtained that showed a patent venous system (Figs. 4 and 5). At the same time, a pulmonary angiogram showed no evidence of large pulmonary emboli. Lytic therapy was avoided because of the patient's hemorrhagic complications after the first re-

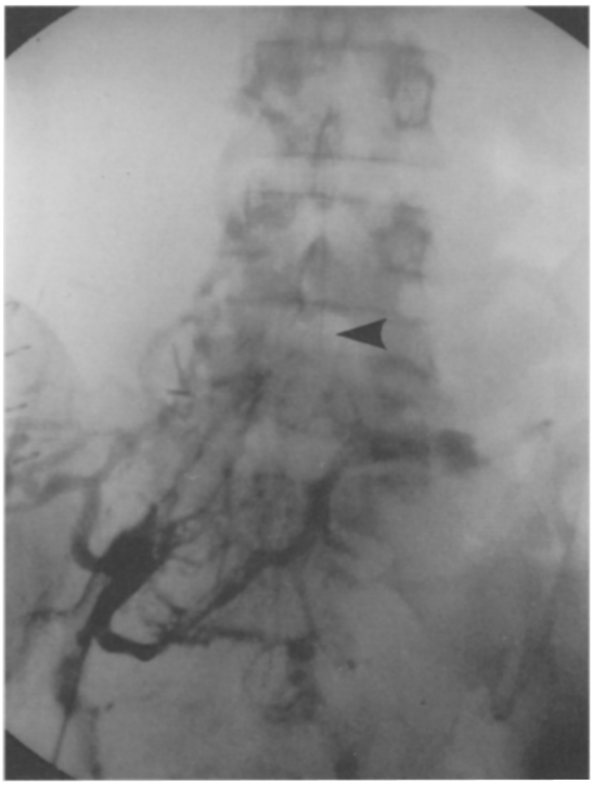

Fig. 3. Rethrombosed inferior vena cava with stent in place *(arrow)*.

Fig. 4. Inferior vena cavagram after recanalization.

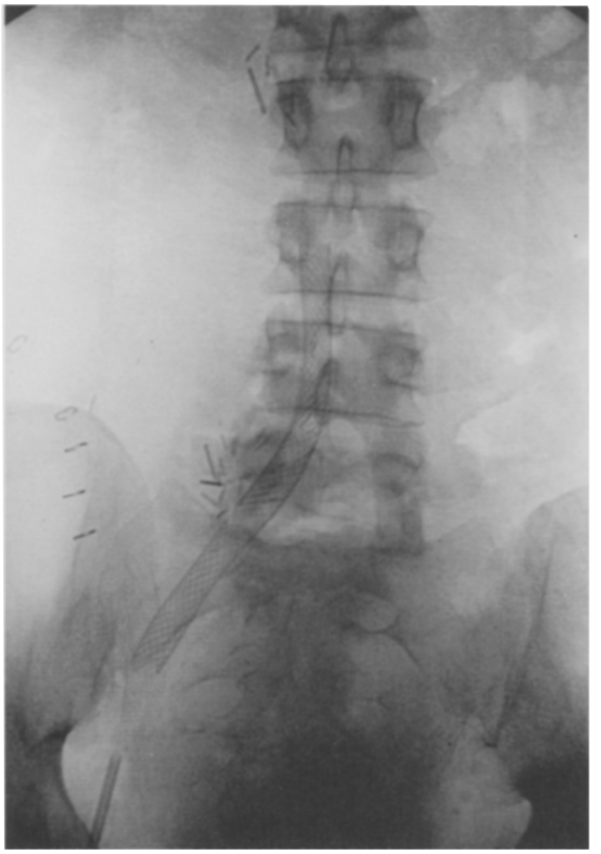

Fig. 5. Final stent positions after recanalization.

canalization surgery. The patient was given heparin in the hospital. She was finally discharged home and resumed treatment with warfarin, with complete resolution of her symptoms. Three months after the procedure, she was able to walk for 20 minutes at 3 miles per hour on a treadmill, demonstrating functional venous return. Eight months after the procedure, a computed tomographic scan with contrast demonstrated a patent venous system (Fig. 6).

DISCUSSION

Of particular interest in this case is the cause of the patient's exercise-induced hypotensive cardiovascular collapse. There are two plausible hypotheses. The first is the increased venous capacitance of the lower extremity caused by the Klippel-Trénaunay syndrome. The alternative is the decreased cardiac venous return caused by vena caval thrombosis. Whether the caval thrombosis was de novo or resulted from the trapping of an embolus is uncertain. Still another possibility represents a contribution of both. It is still unclear how much each disease entity participated in the patient's exercise intolerance and secondary hypotensive cardiovascular collapse.

Gloviczki et al.,[6] in a report of 40 patients with

Fig. 6. Computed tomographic scan with contrast shows Wallstent in patent inferior vena cava *(arrow)*.

Klippel-Trénaunay syndrome, documented that the affected extremity was circumferentially larger in 75% of patients. Furthermore, the authors cited in their series that the greatest difference between extremities was 15 cm, indicating the potential significance of stasis and venous pooling in affected individuals. Venography, though performed in only four patients in Gloviczki's series, revealed significant anomalies of the venous systems in all of the diseased extremities. These included deep venous insufficiency with incompetent perforators, extrinsic compression of the veins, and large dilated deep tibial veins without valves.[6] All such anomalies can cause pooling of the blood supply in the affected extremity and thereby contribute to diminished venous return to the right side of the heart and the resultant exercise intolerance.

Thrombosis of the vena cava, as experienced by the patient in this report, would clearly contribute to decreased cardiac preload. The initial pressure gradient of 17 cm of water across the caval obstruction is verification of this presumption. On recanalization, the pressure gradient was restored to normal and the patient's symptoms were relieved. Furthermore, on rethrombosis of the inferior vena cava, the patient again had symptoms. Subsequent to the recanalization of the stented inferior vena cava, she was again relieved of her recurrent symptoms.

Treatment of vena caval obstruction with stents is a new and innovative technique. Thus far, stenting has mainly been used as a technique to relieve caval obstruction caused by malignancy.[7-10] However, the implications for stenting may encompass a broader spectrum of disease and may promise to be an effective therapeutic alternative to prosthetic or autologous grafts.

Initially, autologous spiral vein grafts were thought to be the optimal method of treatment for obstructed large veins.[11] Doty and Baker[12] and Doty[13] further popularized spiral vein grafts for treating the occluded vena cava. The procedure is tedious, with variable patency rates for spiral vein grafts depending on the location of venous reconstruction. Furthermore, many patients do not have available autogenous vein for grafting.[14,15] Therefore synthetic materials have been used, with the hope of providing an alternative method for venous repair. Expanded polytetrafluoroethylene (ePTFE) and Dacron are among such materials. ePTFE has had the most promising results, both experimentally and clinically.[14-16] It has been reported that a thick pseudointima can develop in the ePTFE graft, thereby reducing the lumenal diameter of the vessel to 60% of its original size.[2] Long-term patency has also been a problem. In view of such complicating factors, stenting may

offer a promising alternative in the treatment of venous occlusion.

The first inferior vena cava self-expanding metallic stent was inserted in 1986.[17] The focus of controversy surrounding the use of the Wallstent has centered on the diameter of the device. There has been speculation that the use of a device with a diameter less than 20 mm cannot be safely inserted into the inferior vena cava for fear of poor wall attatchment.[18] Entwisle et al.[7] recently described three patients who received Wallstents in their inferior venae cavae, all for malignancy associated obstruction. The authors did not report any problems with stent migration, and each of the three patients tolerated the procedure well.[7] The use of stents in the treatment of nonmalignant causes for inferior vena cava obstruction has not been reported thus far in the literature. The patient in this report underwent deployment of a Wallstent to relieve her inferior vena caval obstruction and has had a good result with relief of her symptoms. Three months after recanalization of her inferior vena cava, the stents remain patent by clinical examination, with no evidence of migration by plain abdominal radiograph.

This report describes an unusual patient with vena caval thrombosis and Klippel-Trénaunay syndrome in whom exercise hypotension developed, presumably because of severe disruption of venous return. In correcting her caval obstruction with the use of a Wallstent, an innovative therapeutic technique was used to treat a difficult vascular condition complicated by a rare disorder. Thus far, it seems to have offered promising results.

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